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Three-Dimensional Echocardiography for the Assessment of Atrioventricular Valves in Congenital Heart Disease: Past, Present and Future

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Echocardiography has developed as an imaging technology over 60 years to become the mainstay for investigating heart disease, providing invaluable structural and functional information. In the last 20 years, 3-dimensional echocardiography (3DE) has emerged as an adjunct to 2-dimensional echocardiography in adult and congenital heart disease. Early work with 3-dimensional imaging of the mitral valve describing normal annular shape and function significantly changed the understanding of mitral valve dynamics. Further work led to our current understanding of the mitral valve working as a unit, with all components vital to its normal function. With improving technology and ease of use, similar 3DE techniques have been used in congenital heart disease to study the unique anatomy and function of atrioventricular (AV) valves, specifically the tricuspid valve in hypoplastic left heart syndrome, and the left AV valve in atrioventricular septal defects. This paper describes the role of 3DE in assessing AV valve function in normal valves, and in congenital heart disease.
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Introduction

Echocardiography has evolved from a simple M-mode technique pioneered by Hertz and Edler in the 1950s, to a 2-dimensional modality that revolutionized the diagnosis and management of both adult and congenital heart disease in the 1970s and beyond. The addition of various Doppler techniques then provided additional hemodynamic and functional information. Interestingly, the original work by Hertz and Edler focused on assessment of the mitral valve at a time when rheumatic mitral valve stenosis was at its peak.[1]

In the early 1970s, Dekker and colleagues[2] experimented with 3-dimensional echocardiography (3DE) as a potential application to image the heart using spatial locators.[2] This provided inspiration to other investigators to explore the potential of this new application. Initially the technology was a limiting factor because imaging relied on acquiring a series of slices of the heart and then stitching them back together. Multiple, time-consuming steps were required to produce a 3-dimensional image. The discovery of matrix array technology by Snyder et al[3] changed the playing field. This breakthrough permitted real-time 3-dimensional data to be acquired; this is the technology available today.

Why the need for 3DE when its 2-dimensional counterpart was rapidly becoming the first line of investigation for most forms of congenital and acquired heart disease? We live in a 3-dimensional world and visualizing the heart through a series of 2-dimensional slices does not provide the type of information necessary for an in-depth understanding of a complex anatomical and physiological structure such as the heart. Additionally, 2-dimensional echocardiography (2DE) requires an “interpreter,” the echocardiographer, to provide a description of what the images show. This is particularly relevant to the understanding of atrioventricular valve function, the main topic of this paper. What 2DE shows and what the surgeon sees at the time of the operation are completely different. On the other hand, 3DE can provide precise images of the valves, exactly as the surgeon sees, but with the added advantage of the real-time interaction between the valve and the ventricle throughout the cardiac cycle.

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KEY POINTS

- Two-dimensional echocardiography (2DE) remains the main imaging modality for diagnosis and management of congenital heart disease.
- Three-dimensional echocardiography (3DE) has significantly improved our understanding of atrioventricular valve structure and function.
- In congenital heart disease, 3DE is an important adjunct to 2DE to identify AV valve anatomy and pathology prior to surgical repair.
- Three-dimensional echocardiography may improve communication of AV valve anatomy and pathology to surgical staff.
- Continued evolution of imaging and display technology will improve acquisition, processing and viewing of 3DE imaging.

Current Applications of 3DE

Although the focus of this manuscript is on atrioventricular valve function, there are other applications of 3DE that should be mentioned briefly[4]:

1) Left ventricular function assessment, both global and regional, is one of the main uses of 3DE in adults, providing accurate and repeatable data. Indeed, it is recommended for clinical practice in adults.[4]
2) Measurement of right ventricular volume and function appears promising, and can be obtained in some patients, but this use is currently more experimental.
3) Assessment of aortic valve morphology and function is a relatively new application, but remains challenging. 3DE is good at imaging thickened aortic valves, but is less useful when the leaflets are thin. It is often possible to image the zones of coaptation, but the curved belly of the leaflets can be difficult to image. Inadequate acoustic windows often hamper optimal 3DE imaging of the aortic valve because of the angle at which the aortic valve leaflets sit relative to the plane of the ultrasound beam.
4) Guidance of transcatheter procedures is a common (and recommended) use for 3DE in adults, including atrial and ventricular septal defect closure, transcatheter aortic valve implantation, left atrial appendage closure, and mitral valve intervention (mitral clips or mitral valvuloplasty).
5) Assessment of intracardiac anatomy in complex congenital heart disease is an emerging use for 3DE. Spatial relationships are often complicated, and 3DE imaging can provide a greater appreciation of the anatomy.[5,6] This can be particularly important when planning interventions. In hearts where the systemic ventricle must be tunneled to the aorta through a ventricular septal defect, 3DE can identify complex spatial relationships as well as assess for structures that might obstruct the pathway.[5]

Atrioventricular Valve Function

This is where 3DE has had its greatest impact. Although we are discussing the impact in congenital heart disease, it is important to discuss some of the pioneer work undertaken by Robert Levine, who laid the foundation for insight into mitral valve function.[7,8] His work uniquely addresses the mitral valve as a complex unit with all components interacting in unison with the left ventricle. In two seminal papers, initially he described the saddle-shaped mitral valve annulus, and then the importance of this annular shape for diagnosis of mitral valve prolapse (MVP). These findings radically changed the understanding and diagnosis of MVP, with frequency of MVP now recognized as about 2.4% of the population, rather than the 10% to 15% of earlier years. This relates to the notion that this diagnosis should only be made when the leaflets bulge above the high points of the saddle-shaped annulus (Fig. 1).

Levine also described the relationship between papillary muscle position and annulus. He demonstrated in ischemic mitral regurgitation that the papillary muscle is displaced, with subsequent tethering of the leaflets by the sub-valve apparatus, in particular the strut chords of the anterior leaflet.[9] This work continues, as his group is exploring the genetic basis of MVP and the impact of tethering on structural changes in the leaflets.[10,11]

The Chicago group, under the leadership of Roberto Lang, has also progressed the understanding of mitral valve failure in the adult population, as well as being the catalyst for providing a document on imaging standards on how to acquire and view 3-dimensional images.[4]

Mitral and Tricuspid Valve Interaction

The mitral and tricuspid annuluses are saddle-shaped, with two high points and two low points, to reduce leaflet stress. Changes occur throughout the cardiac cycle to maintain maximum zones of coaptation of the leaflets.[12] The mitral annular function is more heterogeneous than its tricuspid counterpart. This is related to the deep wedging of the aorta and the area of fibrous continuity between the aortic leaflet of the mitral valve and the non-coronary cusp. The heterogeneity permits maximal expansion of the left ventricular outflow tract during systole to permit unimpeded ejection of the blood from the left ventricle.[13,14] The tricuspid valve (TV) annulus is more remote from its outflow tract and has a homogeneous contraction pattern.[15]

The mitral annulus is not as elliptical as the tricuspid annulus, and the interaction between the two is such that the forces are greater in the lateral, rather than the antero-posterior, direction.[15] This is important because, although the septal leaflet of the TV is large, it is relatively immobile compared with the anterior and posterior leaflets, and acts as a “door jam” for the other two leaflets. Conditions that cause the tricuspid annulus to become more circular in shape disrupt this important relationship and potentially lead to TV incompetence.[16]

Mitral and TV Leaflets and Support Apparatus

For years, surgeons have appreciated that the mitral and TVs have peaks and valleys, but only more recently has the